Massive Pulmonary Artery Aneurysm
Causing Left Main Coronary Artery Compression in the Absence of Pulmonary Hypertension

We report the case of a 62-year-old woman who presented with classic symptoms of stable angina. Cardiac images and catheterization results revealed absent pulmonary valve syndrome and compression of the left main coronary artery by a massively dilated pulmonary artery aneurysm. The patient’s anginal symptoms were relieved after pulmonary arterioplasty.

Others have described proximal left main coronary artery compression in the presence of a dilated and hypertensive pulmonary artery. To our knowledge, this is the first case in which a pulmonary artery aneurysm caused left main coronary insufficiency in the absence of pulmonary hypertension—a clinically important complication of congenital pulmonary valve-related pulmonary arteriopathy. (Tex Heart Inst J 2015;42(5):465-7)

In absent pulmonary valve (PV) syndrome, the PV leaflets are hypoplastic and dysmorphic, resulting in severe pulmonary incompetence and (less often) stenosis. Absent PV can occur in isolation, in association with tetralogy of Fallot, and occasionally in association with patent ductus arteriosus and a hypoplastic right heart. The main and branch pulmonary arteries (PAs) are characteristically markedly enlarged and might compress the bronchi or esophagus. We describe the apparently unique case of a patient with this syndrome who presented with exertional angina.

Case Report

In September 2010, a 62-year-old woman presented with a 6-month history of left-sided substernal chest heaviness, which reliably occurred after 20 to 30 minutes of brisk walking on a treadmill or on inclines. The symptoms resolved with rest. The patient reported no respiratory symptoms or dysphagia. Her medical history included a congenitally abnormal PV with an associated massive PA aneurysm. On examination, her vital signs and weight were normal. No findings suggested Noonan syndrome. The patient’s jugular venous pressure was normal, and there were no prominent A waves. Auscultation revealed clear lungs, a normal S1, a single S2, a mid-peaking grade 2/6 systolic ejection murmur over the left upper sternal border with a short grade 3/4 low-pitched diastolic murmur, and no systolic ejection click. There was a palpable right ventricular (RV) lift and no PA pulsation.

An electrocardiogram showed sinus rhythm, incomplete right bundle branch block, precordial T-wave inversions, and a normal QTc interval. An echocardiogram revealed biatrial enlargement, a trileaflet aortic valve without stenosis, hypoplastic and thickened pulmonic leaflets consistent with a congenitally abnormal PV, a peak pulmonic gradient of 41 mmHg (mean, 22 mmHg), severe pulmonic insufficiency, main PA dilation of 7.9 cm, an estimated RV systolic pressure of 38 mmHg (assuming a right atrial pressure of 5 mmHg), severe RV dilation without hypertrophy or dysfunction, mild tricuspid regurgitation, and an intact ventricular septum.

Given the patient’s history of exertional chest discomfort, she underwent cardiac computed tomographic angiography. This revealed compression of the proximal left main coronary artery (LMCA) by the severely enlarged main PA. Immediately distal to the origin of the LMCA was narrowing—with an obliquely oriented, slit-like course, secondary to compression by the massively dilated main PA (Figs. 1–3). The RV end-diastolic volume was severely enlarged at 225 cc/m² (normal, <90 cc/m²). An incidental finding was an anomalous origin of the right coronary artery from...
the noncoronary cusp. Coronary angiograms revealed a 50% tapering stenosis in the proximal third of the LMCA, secondary to compression by the dilated main PA (Fig. 4). Right-sided heart catheterization yielded a right atrial pressure of 6 mmHg, a normal PA pressure of 26/13 mmHg, a pulmonary capillary wedge pressure of 7 mmHg, and a mean transpulmonary valve gradient of 6 mmHg.

The patient underwent surgical PA aneurysm repair by means of branch and main pulmonary arterioplasty.
along with PV replacement with use of a 27-mm bioprosthetic pericardial valve. Of note, upon intraoperative inspection, we found the PV to be trileaflet with rudimentary, thickened, and dysmorphic leaflets. Follow-up computed tomographic angiograms revealed a substantial interval decrease in the size of the PA aneurysm and complete resolution of the LMCA compression (Figs. 5 and 6). The patient recovered uneventfully. As of July 2015, she had no recurrent chest discomfort and reported that she vigorously exercised daily with no functional limitations or chest symptoms.

**Discussion**

Proximal LMCA compression by an enlarged PA was first described in 1957 in patients with severe pulmonary hypertension, both primary and secondary to pulmonary vascular obstruction or Eisenmenger syndrome. Radiographic risk factors for coronary insufficiency include the degree of coronary compression, a left main angle of takeoff from the left sinus of Valsalva of $<30^\circ$, and a main PA/aorta diameter ratio $>2.4$. Some authors consider unprotected LMCA stenting to be a palliative alternative if surgical correction is not an option. Many case reports have described LMCA compression by an enlarged, severely hypertensive PA. Our report illustrates this rare and, to our knowledge, unique occurrence of LMCA compression caused by a massive PA aneurysm in the absence of PA hypertension. Heretofore, coronary insufficiency was not deemed to be a complication of dilated PAs without pulmonary hypertension. This important complication should be considered in patients who have valvular pulmonic stenosis-related pulmonary arteriopathy.

**References**